



Summer vacations not only a memory for life, but also a rare infection.

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Introduction: Rhinoscleroma is a rare, chronic, granulomatous disease that most frequently affects the upper respiratory tract, especially the nasal cavity, and sometimes extends through the lower respiratory tract. It is associated with *Klebsiella rhinoscleromatis*, which is endemic in certain geographic regions namely Central America. The pathogenesis and risk factors remain unclear.

CLINICAL CASE

5 Years old, Portuguese boy

Irrelevant family history

Previously healthy

Summer vacations 2009 Dominican Republic

Emergency Department – July 2010

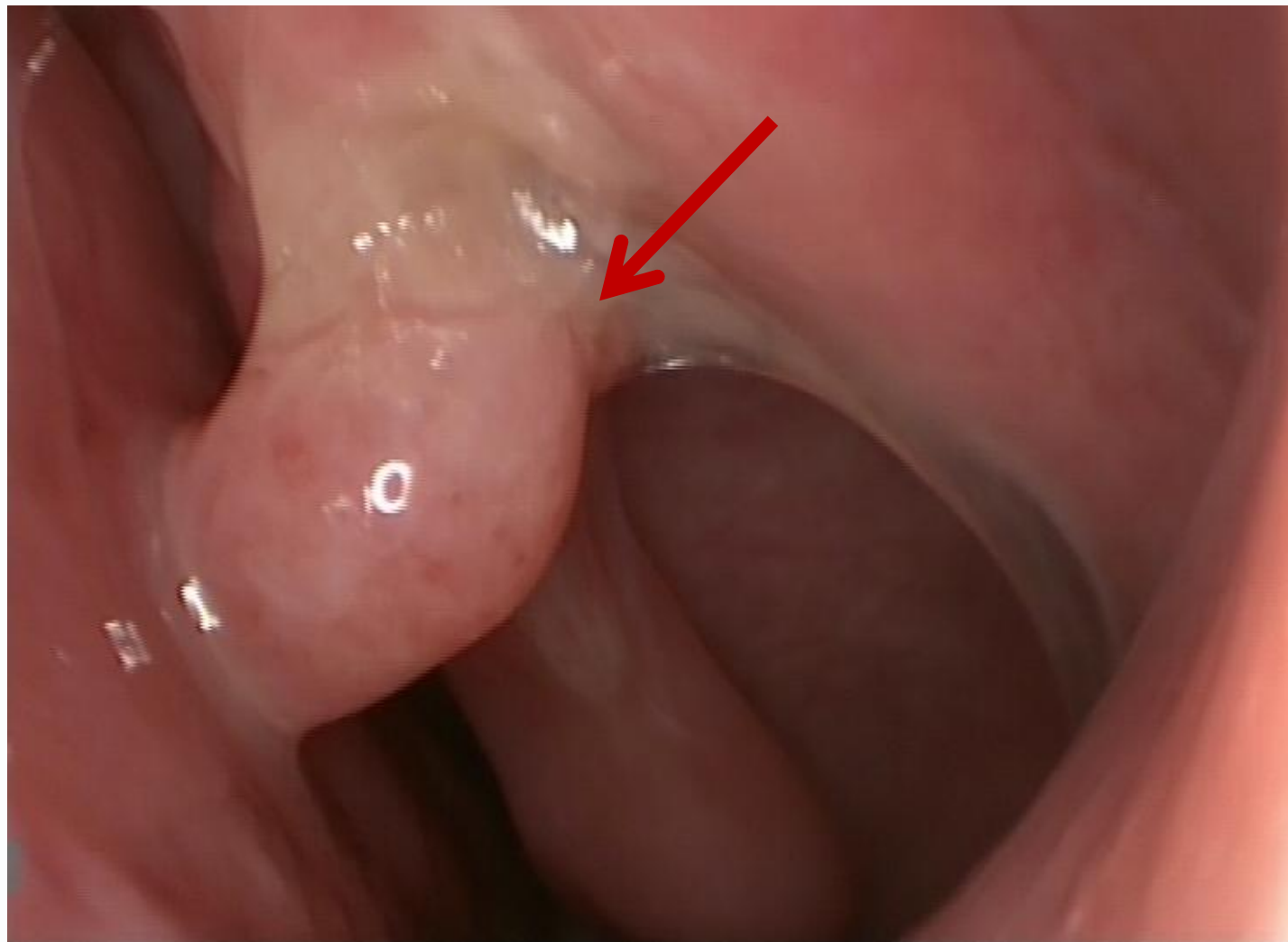
Epistaxis

No other signs or symptoms

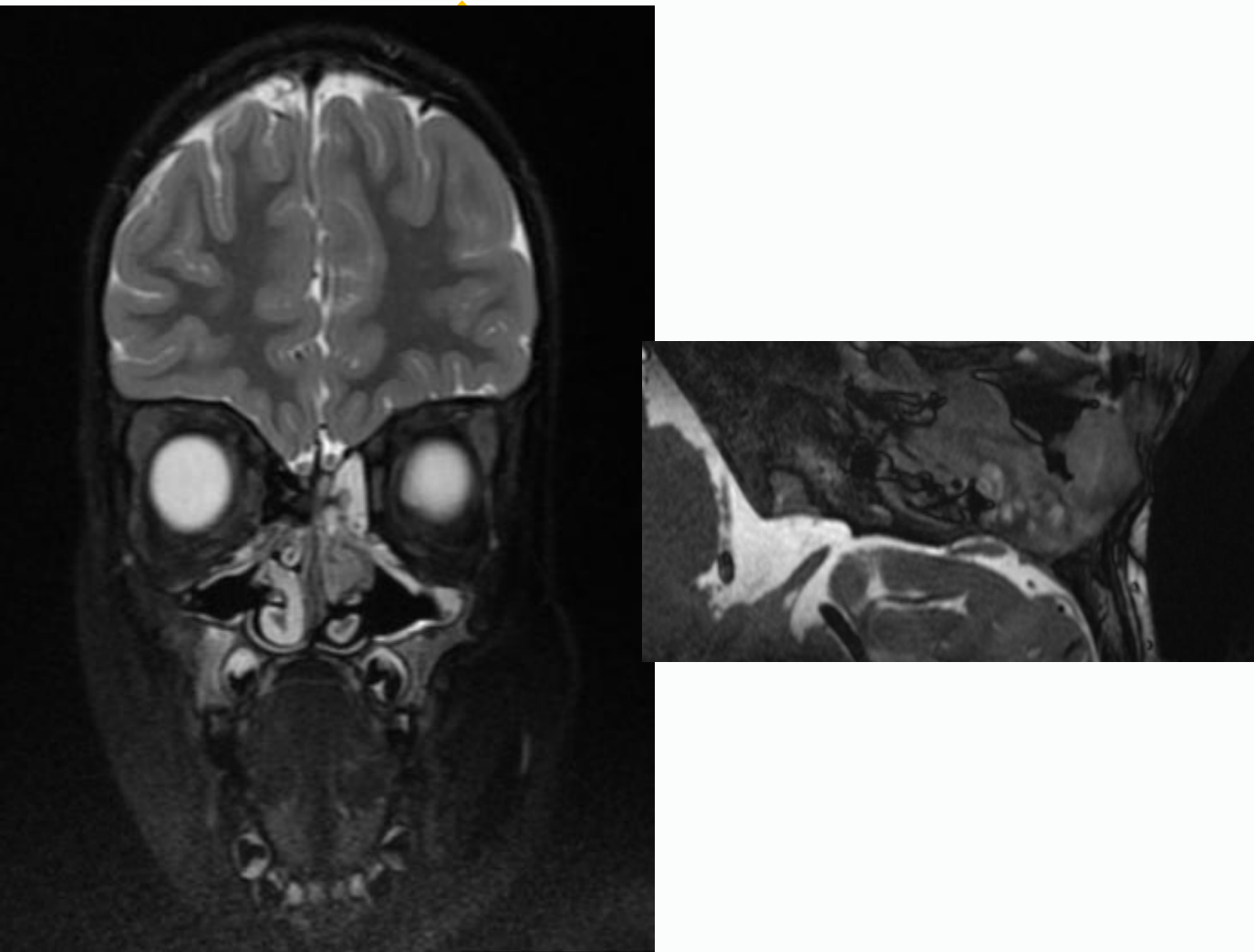
Rhinoscopy **INTRANASAL BLEEDING MASS**

MRI – July 2010

Intranasal mass with extension to ethmoid bone sinus

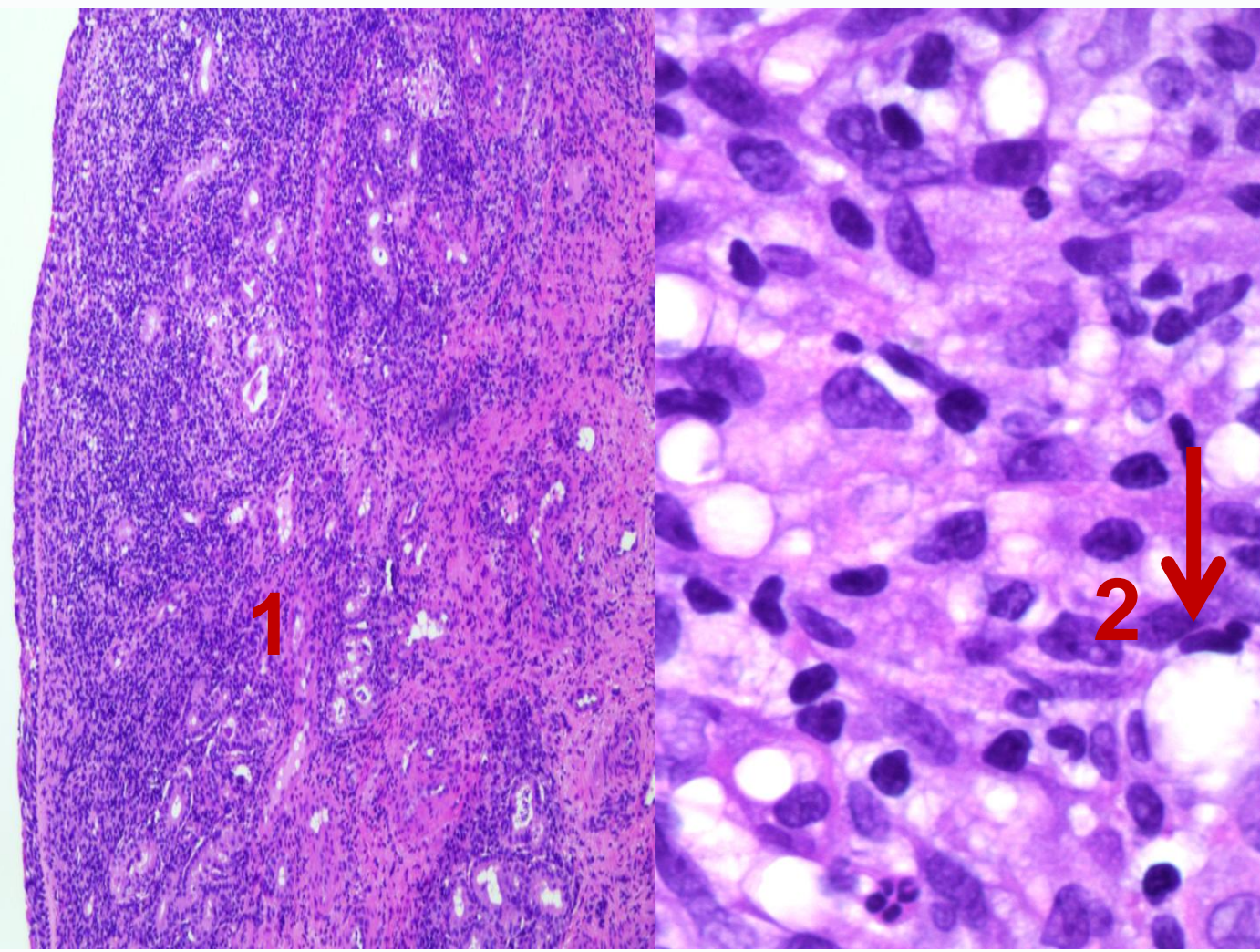


Picture 1 - Rhinoscopy



Picture 2 - MRI

Bacteriologic exam of biopsy material - *Klebsiella Spp.*
sensitive *in vitro* to amoxicillin and clavulanic acid



Picture 3 - Histopathology

HISTOPATHOLOGY PATHOGNOMONIC FEATURE

Granulomas (1) containing
Mikulicz cells (2) - cells with
Klebsiella rhinoscleromatis

RHINOSCLEROMA

GENETIC STUDY

Exome sequencing;
functional study in
immortalized cell lines
Under course

IMMUNODEFICIENCY SCREENING:

Immunoglobulins; Lymphocyte
subpopulations; Phagocytosis test;
Oxidative burst; CH100 **Normal**

Treatment

Amoxicillin plus clavulanic acid

Follow-up

Monthly evaluation - ORL and Pediatrician



Picture 3 - Rhinoscopy

Progressive reduction until
total disappearing of macro
and microscopic lesion
Negative bacteriologic exam

**6 Months of Antibiotic
therapy**

**8 Months After End of
Treatment**

**ASSYMPTOMATIC
No evidence of recurrence**

What is said in the literature!!

- Clinical outcome is variable.
- 3 Stages: catarrhal or atrophic; granulomatous or hypertrophic and sclerotic.
- Antimicrobial therapy must be administered for prolonged periods (3-9 M).
- Needed antibiotics with activity against gram-negative bacilli, intracellular efficacy and low toxicity. Ex: quinolones
- Association with cellular immunodeficiency; weaker antibody responses.
- Genetic predisposition to *Klebsiella rhinoscleromatis* infection may involve a specific pathway.
- Relapse until 3 years.

Comments: Globalization and free transit of people to areas far from origin countries where some rare diseases are endemic brings a new challenge to modern medicine. Sometimes vacations bring more than memories.

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